Introduction to the Single Ventricle Reconstruction trial

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The Single Ventricle Reconstruction (SVR) trial, sponsored by the National Heart, Lung, and Blood Institute and carried out by 15 clinical centers, randomized 549 neonates and showed that transplantation-free survival at 14 months was significantly higher overall with the right ventricle-to-pulmonary artery shunt (RVPAS) compared with the modified Blalock-Taussig shunt (MBTS).[1] Any surgical procedure that does not exceed 65% survival at 1 year deserves to be studied and scrutinized relentlessly. Although the primary outcome was clear when considering the entire study cohort, many questions remained regarding the impact of comorbidities; intraoperative, postoperative, and interstage management strategies on morbidity and mortality; and the mechanisms of death throughout the study period.

The compilation of 5 follow-up articles[2-6] represents secondary analyses of the SVR trial, and their main value is that they provide a more detailed analysis of areas that could not be covered in the *New England Journal of Medicine* article.[1] The SVR trial fortunately included enough study patients, and collected a vast amount of data, to permit analysis of the interaction of risk factors with shunt type[2]; the risk factors of hospital morbidity and mortality after the Norwood[3]; the impact of shunt type and other risk factors on interstage mortality[4]; the cause, timing, and location of death after the Norwood[5]; as well as a descriptive study on practice variation in perioperative care found across centers.[6]

Some of the findings confirm the findings of previous retrospective studies or database extraction studies, such as the impact of center/surgeon volume, the major and independent role of extracardiac anomalies, genetic syndromes, low gestational age and birth weight, smaller ascending aorta, obstructed pulmonary venous return, and lower socioeconomic status play on outcomes. Patients with aortic stenosis/mitral stenosis did better than any other subgroup, presumably because of the presence of antegrade aortic flow, and patients with aortic atresia/mitral stenosis did not exhibit a higher mortality, although the presence or absence of coronary fistulae was not documented. Another novel and noteworthy finding of these secondary analyses include the observation that term infants with aortic atresia (51% of the cohort) did considerably better with an RVPAS at 12 months (mortality one third that of a similar patient with MBTS), which is significant and lines up neatly with a physiologic explanation of superior diastolic coronary flow and lack of coronary steal in RVPAS patients. Conversely, in 4% of the cohort (preterm infants with a patent aortic valve), the MBTS was superior. No clear explanation exists for this finding, and one could speculate that the ventriculotomy is less well tolerated in preterm infants. This was a much smaller group of patients and thus statistical inferences are more suspect. For term infants with a patent aortic valve and for preterm infants with aortic atresia, no clear difference between RVPAS and MBTS was seen. This may be because the former group does well with either shunt type and that, in the latter group, the benefit of the RVPAS on coronary flow may be offset by the downsides of a ventriculotomy.

Open sternum was identified as a significant risk factor independent of the center's strategy (elective vs routine sternal closure). However, the study did show that, in selected patients, primary sternal closure is a safe alternative. These types of associations, however, are always difficult to interpret because this ''outcome variable'' is dependent on the surgeon's decision at the end of the procedure and may reflect patient instability or even a concern with technical aspects of the surgery that are not captured by the usual data-gathering process.

Other notable findings were that interstage mortality was significantly better for the RVPAS compared with MBTS, unless atrioventricular valve regurgitation was moderate to severe, in which case this overshadowed the shunt type and interstage mortality was similar. Not surprisingly, most deaths occurred during initial hospitalization. The nonspecific determination of ''cardiovascular death'' was the most common cause, a sure way to frustrate readers who will long to get some clues as to what often remains an unexplained phenomenon: the sudden unexplained death after Norwood. It is important to note that whatever autopsy data were available were in large part nonrevelatory.

In conclusion, these combined studies point to the RVPAS as being beneficial in term infants with aortic atresia and as being associated with lower interstage mortality in general. For term patients with aortic stenosis/mitral stenosis, either shunt is associated with better survival. These studies represent significant new knowledge. Sadly, improved survival after Norwood, let alone quality of life,

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Disclosures: Authors have nothing to disclose with regard to commercial support.

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J Thorac Cardiovasc Surg 2012;144:880-1
0022-5223/$36.00

http://dx.doi.org/10.1016/j.jtcvs.2012.08.031

remains an elusive goal that we as a profession are reaching only incrementally.

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